Prosthodontics rehabilitation of a patient with hypohidrotic ectodermal dysplasia with partial anodontia: Case report

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Abstract

Ectodermal dysplasia (ED) belongs to a large group of heterogeneous genetic disorders that is characterized by tissue dysplasia of ectodermal origin and commonly involves the skin, hair, nails, eccrine glands and teeth; although, at times it may be of mesodermal origin. As is often found, patients with this disorder present with nail dystrophy (onychodysplasia), alopecia, or scanty, fine, very light hair on the scalp and eyebrows (hypotrichosis), and palmoplantar hypohidrosis, which is commonly accompanied with lack of sweat glands and complete or partial primary and permanent dentition. Ectodermal dysplasia usually manifests as X-linked recessive hypohidrotic ectodermal dysplasia (HED) and has a full expression in males, while females show little to no signs of the disorder. This case report discusses the management of a young boy with hypohidrotic ectodermal dysplasia that was treated with removable prostheses. The aim was to prosthodontically rehabilitate the child and help him psychologically.

Key words: Hypohidrotic ectodermal dysplasia, Partial anodontia, Prosthetic therapy

Introduction

Ectodermal dysplasias (ED) are congenital disorders that are described as large and heterogeneous and were initially described by Thurnam in 1848 and then Darwin in 1875. ED was assigned to the X chromosome in 1921 by Thadani, who later reported that carrier females could manifest signs of the condition. The occurrences are relatively rare (1/100000 births) and it is characterized by the abnormal development of the ectodermal

derivatives.[1,2,3] Oral health findings include total or partial anodontia affecting both the primary and the permanent dentitions. The teeth are usually conical or peg shaped. Alveolar ridges are underdeveloped resulting in reduced vertical dimension, thereby giving a senile facial appearance along with protuberant dry lips.[2,4,5] This paper describes the prosthetic rehabilitation of a child with ED presenting severely resorbed alveolar ridges.

Case Report

A 7-year-old boy presented with a complaint of missing teeth. According to his parents, since infancy he suffered from missing teeth, except for two deciduous teeth in the upper jaw. The parents also gave a history of intermittent fever throughout infancy and childhood. There was no family history of missing teeth. The boy was moderately built and well nourished. His skin was extremely dry and he hardly had any body hair. His facial features were common of an ED sufferer such as periorbital pigmentation, frontal bossing, a depressed nasal bridge, protuberant lips, low-set ears, and scanty scalp hair.[2,6] Nails were not very brittle and an intra-oral examination exposed a relatively dry mucosa. The mandibular arch was fully edentulous with poorly developed alveolus and the maxillary arch had two standing permanent molars [Figure 1] and [Figure 2]. Since hypohidrosis, hypotrichosis, and hypodontia were evident in the physical examination, the boy was diagnosed with Hypohidrotic Ectodermal Dysplasia (HED).

Prosthodontic Management

The prosthodontic management of children suffering from HED depends on the degree of anodontia/hypodontia. In complete anodontia, the treatment would comprise of complete dentures, either conventional or implant supported ones.[7] In patients with partial anodontia, removable/fixed partial dentures and over dentures may be considered. In the present case, a removable partial denture in the maxillary arch and a conventional complete denture in the mandible arch were selected. Preliminary impressions were made using alginate (Alginoplast fast set, Heraeus-Kulzer, Holland). Special trays were fabricated using autopolymerizing acrylic resin (MelioDent, Heraeus-Kulzer, Holland) after applying spacers (Modeling wax, MDM Corp, USA) to relieve the mid-palatine raphe, incisive papilla, and the standing molars in the maxillary arch and the crest of the ridge in the mandible arches. Border molding was done using a green stick compound (Harvard Impression Compound, Harvard Dental, Germany). Secondary impressions were made using the Zinc Oxide Eugenol Impression (ZnOE) paste (Impression Paste, DPI, Chicago, USA) for the mandible arch and light viscosity addition silicone (Reprosil, Dentsply, USA) for the maxillary arch [Figure 3] and [Figure 4]. Record bases (cold cure acrylic resin, DPI and modeling wax, MDM Corp, USA) were made and jaw relations registered. The selection of teeth for children and adolescents requires more attention as the commercially available moulds do not satisfy esthetic requirements in most cases; obviously because these moulds mimic adult dentitions.[8]

A vigorous-looking mould was chosen to provide a masculine appearance (Major dent, Major, Italy). But considering the age of the boy and the soft facial features common in a boy his age, we tried to tone down the masculine-looking mould by arranging the maxillary anteriors to mimic a C 1 II, division II case. Very mild crowding was replicated in the mandibular anteriors to enhance a natural appearance. Posterior teeth were arranged according to the boy's natural teeth. Trial dentures were checked for retention, phonetics, occlusion, and esthetics. Acrylic dentures were processed (DPI Heat Cure Acrylic Resin, DPI) through compression molding and the polished dentures were inserted [Figure 5] and [Figure 6]. The boy was taught how to insert and remove his dentures and was given post insertion instructions on denture wear, functions of speech, mastication, hygiene, and maintenance. We asked him to have a liquid/semi-solid diet until he was completely at ease while eating with the dentures. We explained and emphasized the importance of the existing deciduous maxillary molars and the need to maintain them. He was recalled 24 hours after denture insertion for his first post insertion checkup. Later, he was kept on monthly recall visits. He reported inconvenience in mastication for more than two weeks even with a semisolid diet. But the esthetic satisfaction was so immense that he wore the dentures and willingly adapted to masticating and speaking with them.

Psychological aspects

At his initial visit, the young boy was clearly timid and had a difficult time communicating. As a result, he relied on his father to answer most of our questions. Thus, we tried to make him feel comfortable in communicating with us by asking him simple yes-no questions. When we noticed he started to become a bit comfortable, we began open ended questions that required longer responses. Consequently, we understood that his lack of communicate skills were due to his condition and not lacking of intelligence. We noticed an obvious change in his demeanor after his treatment and during his recalls; to one of happiness and ease at communicating. Also, because of this positive change in his life he was notably careful with his dentures and oral hygiene.

Discussion

Oral rehabilitation of an ED patient is needed to improve the sagittal and vertical skeletal relationship during craniofacial growth and development, as well as to provide improvements in esthetics, speech, and masticatory efficiency.[12] Patients with craniofacial anomalies should have a treatment plan that includes not only the management of the defect or anomaly itself, but also the psychological management of the person.[9,10] So, the treatment plan of a child suffering from HED should always include prosthodontic and psychological aspects.[11] Although removable complete or partial dentures are still the preferred treatment, clinicians should consider the use of FPDs and implant-supported prostheses. Children may suffer from severe midface hypoplasia that would require early growth modification and implants; however, implants

are largely indicated in cases with complete anodontia. Obviously, dentures cannot whole replace healthy dentition, but they can create the right circumstances for the establishment of a normal, healthy daily diet that can last a lifetime. Also, in the absence of occlusal stops (or dentures), the anterorotation of the mandible causes an upward and forward displacement of the chin, with a reduction in the height of the lower-third of the face; a tendency to C1 III malocclusion. Dentures help to position the chin.[12]

Periodic recalls are often necessary in young patients as a result of continuing growth and development that mandates adjustments in the prostheses.[13]

Early rehabilitation of children with ED will go a long way in helping them interact normally and integrate them with their peers. Restoration or prosthesis should offer dentition that match the age of the patient.

Conclusion

Early prosthetic therapy for children with ED provides a unique opportunity for a cooperative effort between the pedodontist, periodontist and the prosthodontist; as pedodontists are better trained in the psychological management of children. Prosthodontic treatment may begin at an early age of 3-4 years as a condition for growth and development of orofacial structures. It helps normalize the function of masticatory and perioral muscles and as a result, the growth pattern of basal bones.[14,15]

Conflict of Interest: None declared.

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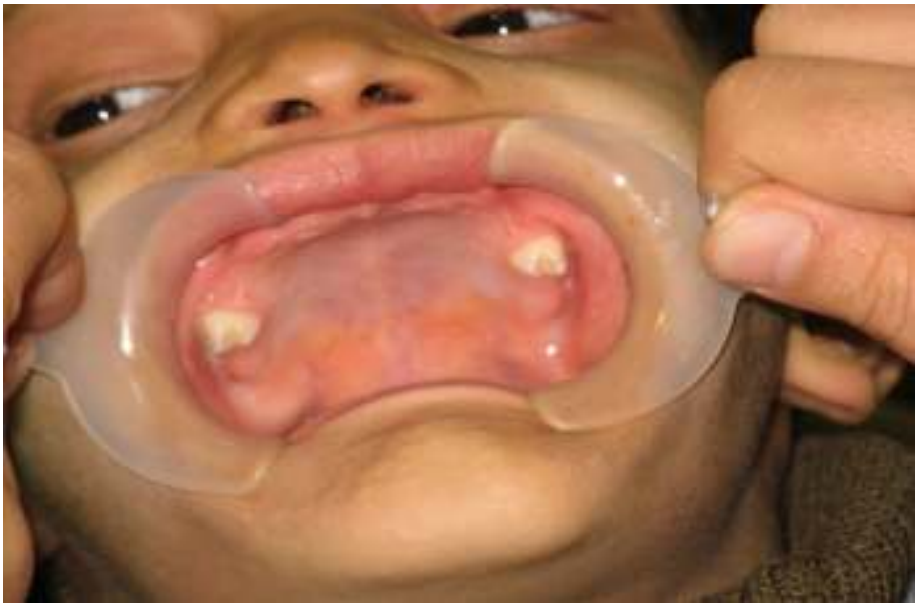

Figure 1: Maxillary arch with two standing deciduous (severe hypodontia)

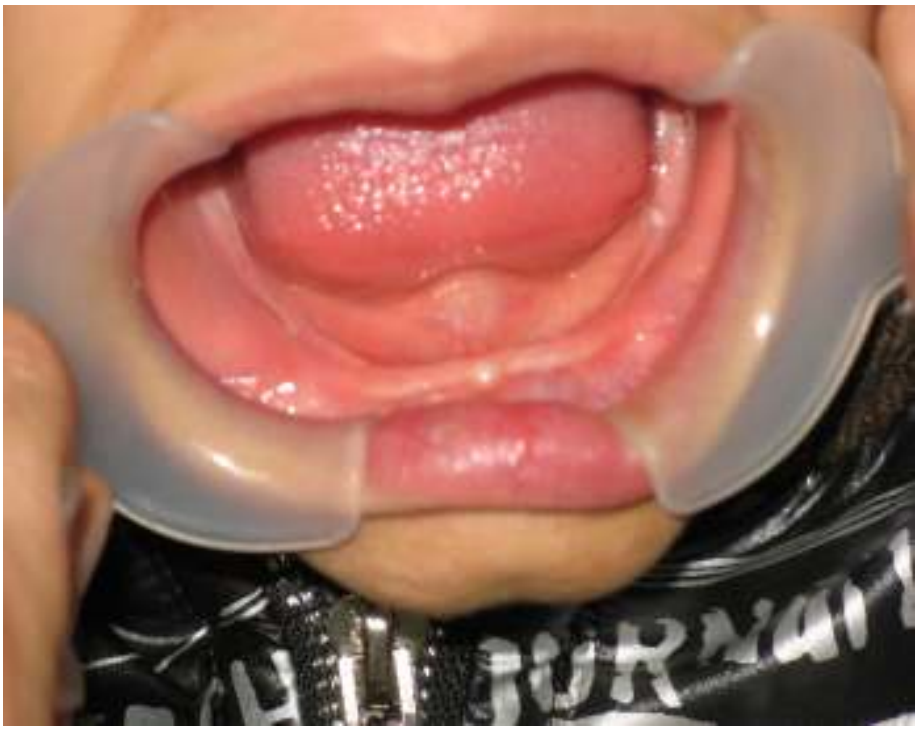

Figure 2: Shows intra oral Mandibular arch – completely edentulous

Figure 3

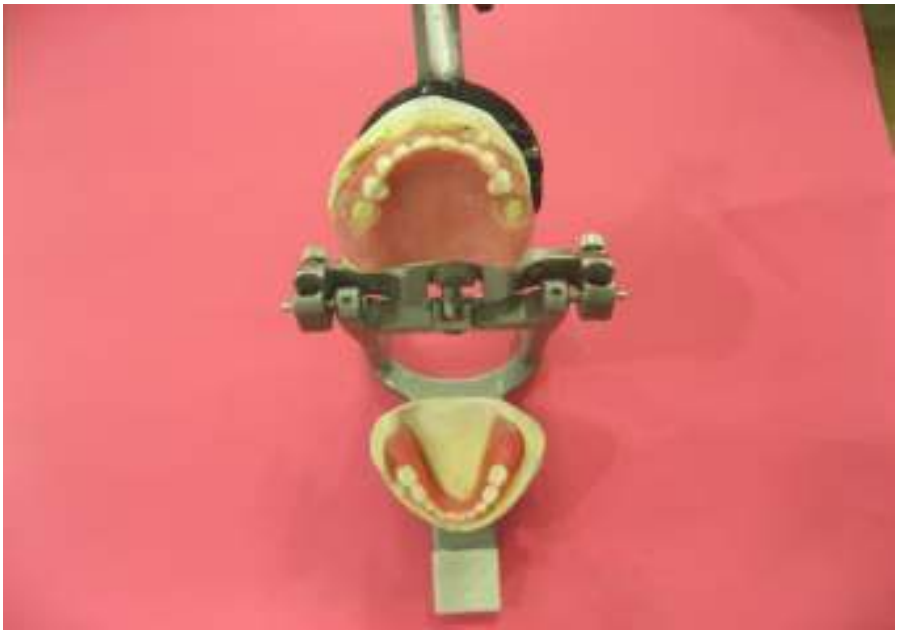

Figure 4

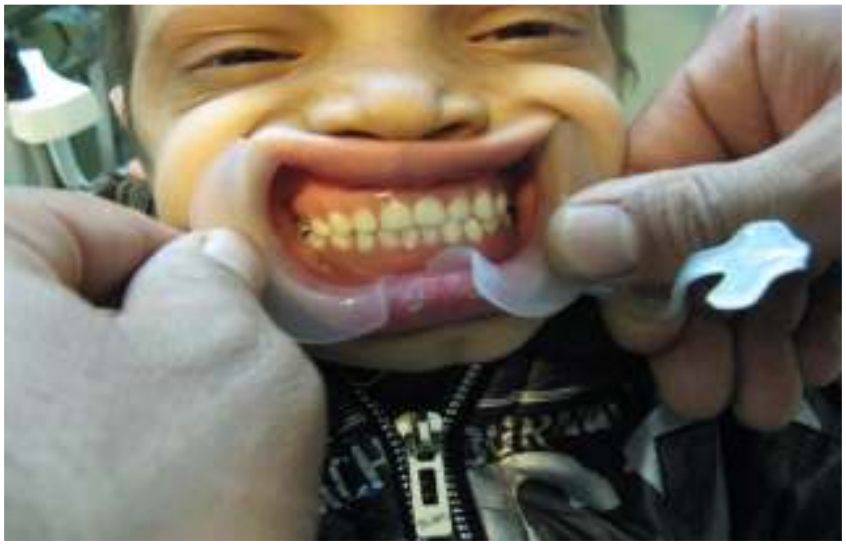

Figure 5: Intraoral view of dentures in occlusion

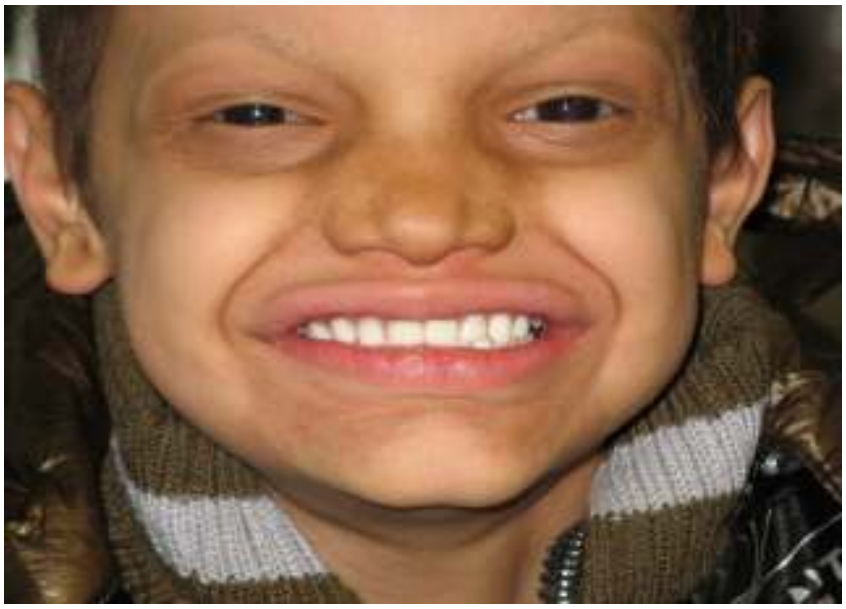

Figure 6: A confident smiles with dentures in place